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症 例 報 告

TUMOR OF THE SYMPATHETIC NERVOUS SYSTEM A CASE OF GANGLIONEUROBLASTOMA

by

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Tumors developing from the sympathetic nervous system include the sympathogonioma, sympathoblastoma, ganglioneuroma, and the paraganglioma. These tumors grow from various parts of the body. Some arise in the adrenal gland, but others grow from the sympathetic tissue anywhere along the retroperitoneal, retropleural, or cervical spaces.

In this paper, the following case of the ganglioneuroma with some areas of neuroblastoma situated in the retroperitoneal space is reported.

CASE REPORT

E. T., a 24-month-old female infant, was admitted to surgical clinic of our Hospital on Apr. 15, 1957, complaining of meteorism.

The abnormal distension of the abdomen was noticed already since she was born, and it became progressively larger day after day. On admission it was huge and more remarkable in the right flank. No abdominal pain, nausea, vomiting and jaundice had been complained. Stools had been normal in color and frequency. Just before the admission there appeared pollakisuria, without haematuria and any other urinary symptoms.

The family history revealed the evidence of asthma and apoplexy.

PHYSICAL EXAMINATION : Examination revealed a moderately nourished obese infant and did not appear acutely ill. The temperature was normal, the pulse rate was 128 per minute, and the respiratory rate was 23 per minute. The blood pressure was 118/72 mmHg. No enlargement of lymph nodules. Neither swellings nor deformities were found in the skull and extremities. Exophthalmus, ecchymosis or edema of eyelids were not proved. The heart was normal in size and rhythm. No murmurs were heard. The lungs were normal by percussion and auscultation.

Positive physical findings were limited to the abdomen. A diffuse and striking distension of the abdomen was observed especially in the mesogastrium. The dilated veins of the abdominal wall ran from the right flank to the right

inguinal region, but not forming *caput medusae*. Peristalsis was active but not violent.

On palpation, the abdominal wall was soft, and neither signs of peritonitis nor ascites were recognizable. In the right flank, there was a tumor having following characteristics : as large as child's head in size, with smooth surface, firm elastic consistency, clear borders and no tenderness. The mass extended upward to the right costal margin, medially to approximately 2 cm right from the midline and downward to the anterior superior iliac spine. The borders were marked as the Fig. 1. The tumor could be grasped bimanually, but showed no syncronous movement with respiration, nor could be movable passively. The sharp and soft edge of the liver was palpable at about 5 cm under the right costal margin at the mammillary line. The surface of it was smooth and there was no tenderness.

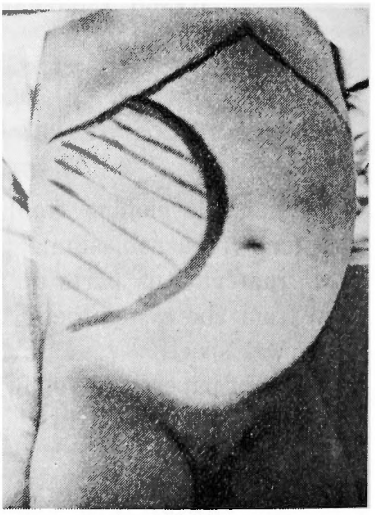


Fig. 1. Preoperative photograph of abdomen with diffuse and striking distension. Markings on the skin show outline of the tumor.

On percussion of the abdomen, the area covering the tumor was dull and the other parts tympanitic. Similarly, the right side in the back was entirely dull.

Pelvic and rectal examinations revealed no abnormalities. Normal brown stool was present in the rectum.

The urine was yellow, alkaline in reaction, with a specific gravity of 1.020. Numerous red blood cells and coli bacilli were found in the urinary sediment. Albumin and urobilinogen was present in urine.

The hemoglobin content in blood was 76%. The red blood cell count was 5.2 millions. The white blood cell count was 5,200, with a differential count of 71% polymorphonuclear leucocytes, 24% lymphocytes, 4% eosinophils and 1% monocytes. The bleeding time was 2 min. 15 sec. Occult blood test of feces was negative.

The examination of hepatic function revealed the positive result in the cobalt reaction, and the other serum turbidity tests were negative.

ROENTGENOLOGIC EXAMINATION : On the plain film study, the borders of the tumor were clear as the Fig. 2. Neither calcification nor abnormally opaque and radiolucent areas were noted in the mass.

Barium enema showed no intrinsic abnormality of the large bowel, but the hepatic flexura was displaced medially, and the ascending colon was compressed by the tumor (Fig. 2).

Excretory pyelography was performed, by means of intravenous injection of Urografin (SCHERING A. G.). The pyelogram showed the lateral and upward displacement of the right kidney by the tumor. The renal outline has been maintained. In addition to those findings, there was abnormal pyelocalyceal pattern of

the right kidney, i e caliectasis and pyelectasis, indicating the hydronephrosis which had been occurred by compression to the right ureter (Fig. 3). On the left side, the normal shadows of the renal pelvis and ureter were appeared. By double printing of photograph 2 and 3, we could understand the relationship of tumor, kidney and colon, more clearly (Fig. 4).

Extrarenal retroperitoneal tumor, such as ganglioneuroma, lipoma, or teratoma was considered as the preoperative diagnosis.

OPERATION : The operative procedure was carried out on Apr. 20, 1957, under drop ether anesthesia. An anterior

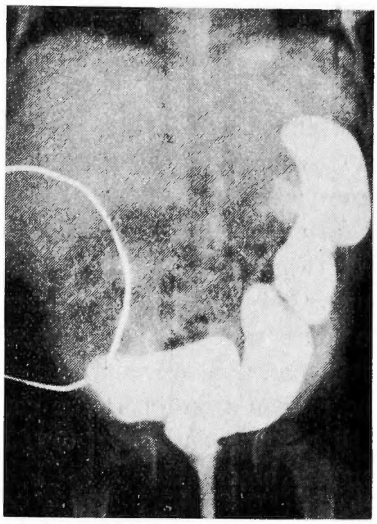


Fig. 2. Barium enema before operation. Film markings show the border of tumor. Neither calcification nor radiolucent areas noted in tumor.

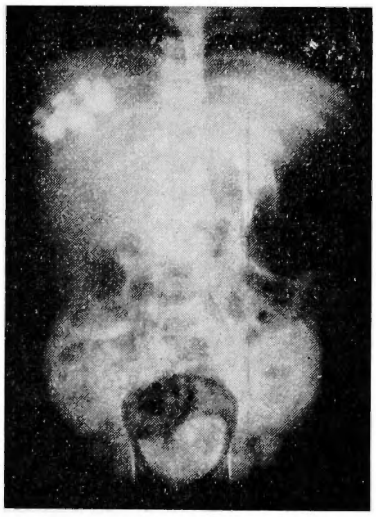


Fig. 3. Intravenous pyelogram shows upward displacement of right kidney. Caliectasis and pyelectasis are noticeable.

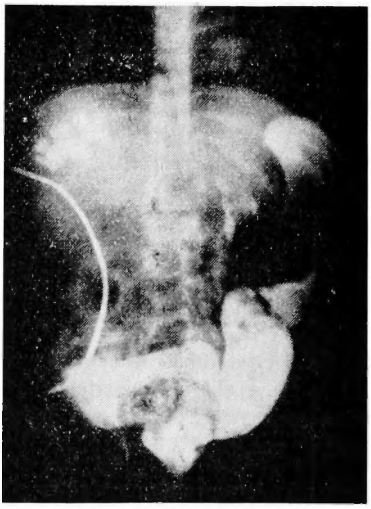


Fig. 4. Double printing of Fig. 2 and Fig. 3, shows the relationship of tumor, kidney and colon.

right-angled incision was made, an extraperitoneal approach being used. Medial retraction of the ascending colon disclosed a huge tumor situated in retroperitoneum. The tumor was as large as a child's head in size, spherical in shape, reddish-white, nodular or partially smooth, firm elastic, scanty in blood vessels on the surface and encapsulated by a fibrous thick membrane. Over the tumor, the inferior vena cava and the right ureter passed, and they were adherent firmly to the mass. Therefore, the bleeding from the inferior vena cava and its branches occurred many times

when the vessel was separated from the tumor. The right kidney was displaced upward, compressed strikingly, and situated between the liver and the tumor. The tumor arose from the right lumbar sympathetic chain and at the site of origin it twined around the vertebra and the abdominal aorta, adhering to them. Furthermore it was partially buried in the right ilio-psoas muscle. Because of this anatomical complexity, a bit of mass remained unremoved at its bottom in spite of careful attempt for the complete removal. The abdomen was closed without drainage (Fig. 5).

During the operation, at times the patient became serious, but she withstood the procedure well by the appropriate blood transfusion sometimes given directly into the vena cava, oxygen inhalation and administration of the cardiant.

The patient's postoperative course was uneventful, and she was discharged of her 16 th hospital day (Fig. 6). Now five months after the operation, no sign of recurrence is observable.

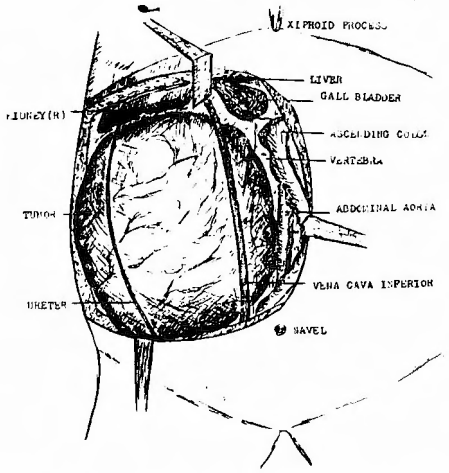


Fig. 5. Schematic illustration of the tumor situated in the right retroperitoneal space. The colon was retracted to the left, the huge tumor was exposed.

Fig. 6. Anterior right-angled wound two weeks after operation.

PATHOLOGICAL EXAMINATION : Macroscopic findings : The specimen was a mass of tissue weighing 700 gm and measuring $14 \times 11 \times 10$ cm. The external surface had a yellowish-red color, and was nodular and partially smooth. The mass was parenchymatous tumor having firm elastic consistency (Fig. 7). The cut surface of the tumor was reddish-white, and showed the appearance of fibrous or connective tissue without showing necrosis and hemorrhage (Fig. 8).

Microscopic findings : The histological specimens taken from the several parts of the tumor were provided to stain with usual haematoxylin-eosin staining and special method such as BIELSCHOWSKY's silver impregnating, EHRLICH's acid hematoxylin, and VAN GIESON's acid fuchsin method.

In the hematoxylin-eosin preparation, the tumor cells showed marked variation

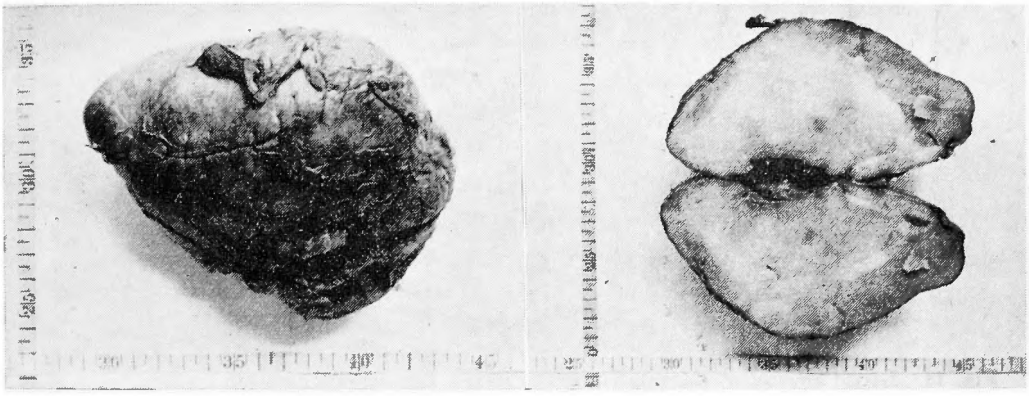


Fig. 7. Anterior view of gross specimen.
Fig. 8. Appearance of the specimen after hemisection. Note the parenchymatous tissue.

in size, shape, and intensity of staining. The cellular components of the tumor consisted of sympathogonia, sympathoblast, and ganglion cell. The small round dark cells resembling lymphocytes were considered to be the sympathogonia. The sympathoblast was characterized by the size which was two or three times as large as the former, and the round shape. The cell nuclei had a spherical contour, and a moderate amount of chromatin. The nucleoli were seen. The cytoplasms were scanty, clear and poorly stained. The main cell components were the differentiated ganglion cells. They were larger, oval or irregular in shape, with 1, 2 or more elongated cytoplasmic processes. Above mentioned three kinds of cells were arranged irregularly. Rosette or pseudorosette formation was not proved (Fig. 9).

Observing the silver stained preparation, we recognized a number of nerve fibers running in every direction. In addition, the Schwann cells were noticeable in the fibrous stroma. Intercellular neurofibrils forming a fine network also could be seen (Fig. 10, 11, 12).

In the acid-hematoxylin staining preparation, non-myelinated bundles of nerve fibers were identified, in which only a few myelinated nerve fibers were found (Fig. 13).

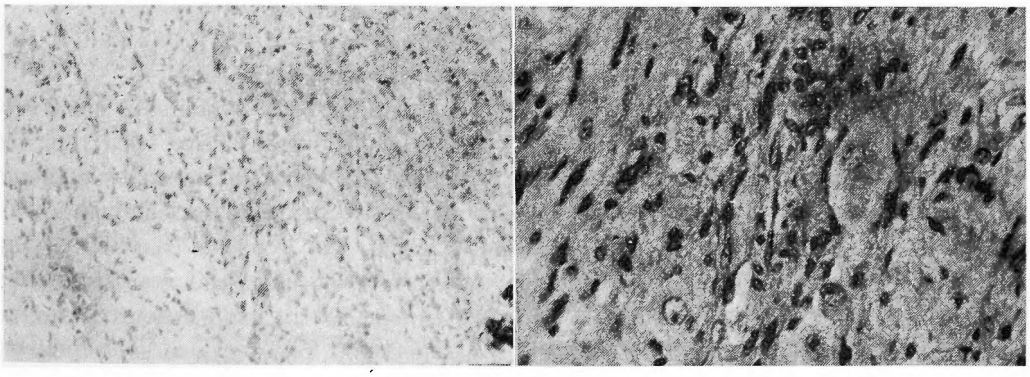


Fig. 9. Showing large ganglion cells, small round dark sympathogonia, and round poorly stained sympathoblast. Hematoxylin-eosin stain, Left (×70), Right (×280).

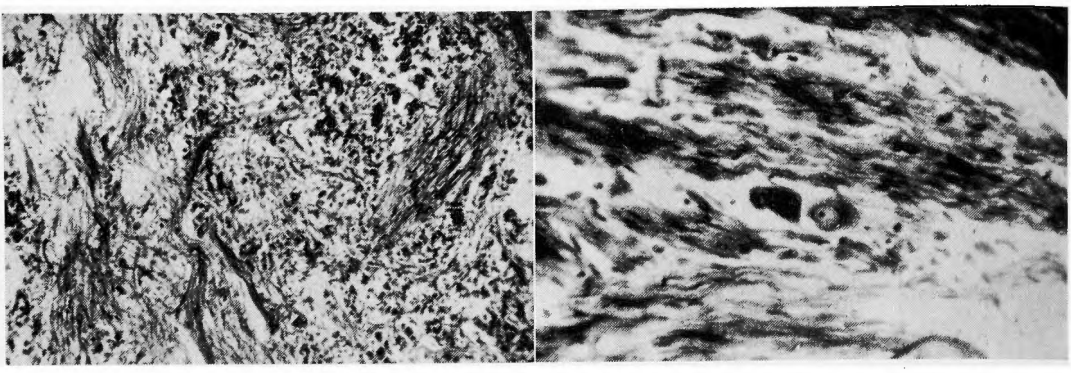


Fig. 10. Large amount of argentaffin nerve fibers. BIELSCHOWSKY's stain (×70).
Fig. 11. Typical ganglion cell. Schwann cells are also seen. BIELSCHOWSKY's stain (×280).

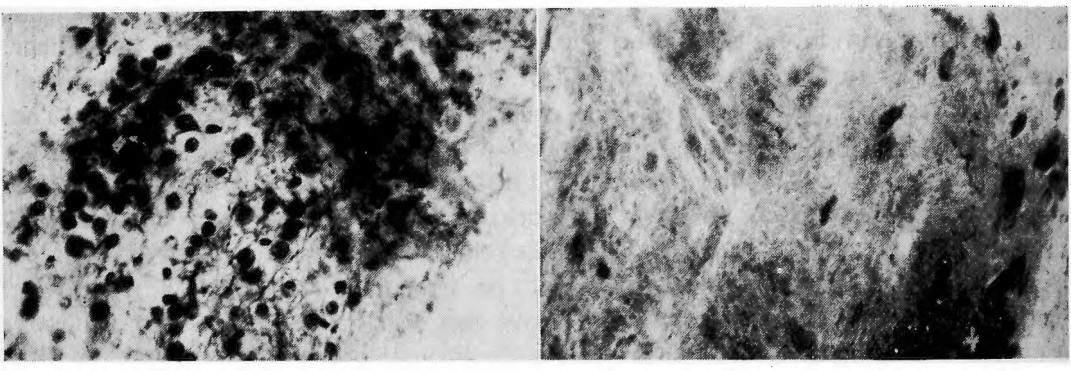


Fig. 12. Showing a number of sympathoblasts and intercellular neurofibrils. BIELSCHOWSKY'S stain (×280).
Fig. 13. A few myelinated nerve fibers are seen. EHRLICH'S stain (×280).

By the VAN GIESON's acid fuchsin method, it was confirmed that the interstitial components were not collagen fibers but nerve elements.

Histopathological diagnosis was a ganglioneuroblastoma.

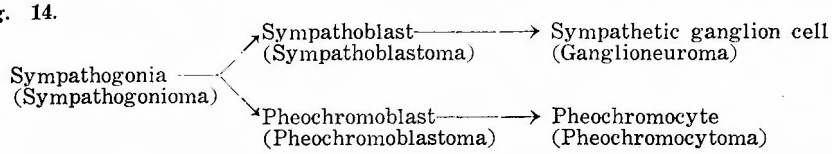
DISCUSSION

The occurrence of tumor from the sympathetic nervous system is relatively rare comparing with other neurogenic growth. About 60 cases have been reported in Japan by KOJIMA and other workers, since the first report of sympathogonioma was described by FUJIYOSHI in 1909. In foreign literatures, the first description of ganglioneuroma was published in 1870 by LORETZ, who reported a case of an encapsulated growth containing wellformed unipolar nerve cells and non-medullated nerve fibers. Since his description, the cases of tumors which include neuroblastoma, ganglioneuroma, and paraganglioma have been reported. The total cases which we find in the literatures are over 300 cases.

PATHOLOGY : The tumor of the sympathetic nervous system is classified

on the basis of histogenesis as Fig. 14 by HERXHEIMER, LANDAN and BIELSCHOWSKY.

Fig. 14.



As ANDERSON describes, the most immature cell forms, the precursors of the other types, are the sympathogonia, which differentiate into sympathoblasts and pheochromoblasts. The sympathoblasts mature as ganglion cells, whereas the pheochromoblasts develop into pheochromocytes.

Tumors composed of the immature cells are called sympathogonioma, sympathoblastoma, and pheochromoblastoma, respectively. Particularly, en bloc sympathogonioma and sympathoblastoma, the term of neuroblastoma is used. Tumors composed of the mature cells are ganglioneuroma and pheochromocytoma. When a tumor is composed of both the immature and mature cells, it is designated as a ganglioneuroblastoma, neuroblastoma-ganglineuroma, neuroblastoma differentiating into ganglioneuroma, well or less well differentiated ganglioneuroma, ganglioneuroma with some areas of neuroblastoma, and etc. Our case was a ganglioneuroblstoma. The character of each cell was described already in the part of case report. The detail of pheochromocytoma is not mentioned in this paper.

SITES OF ORIGIN : Sympathetic nerve tissue and adrenal medulla have the same origin from ectoderm in embryonic development. Therefore, the tumors grow in about the same frequency in the adrenal gland, the abdominal and the thoracic sympathetic chain. We obtained the following statistics from a review of the literatures reported by SUZUE and the others in Japan : adrenal medulla origin; 8 cases, abdominal sympathetic chain origin ; 2 cases, thoracic sympathetic chain origin ; 5 cases. A case in this paper arose from the right lumbar sympathetic chain.

AGE AND SEX : The majority of neuroblastomas reported by SAKAI and the others occurred before the age of four years. LEWIS and GESCHICKTER reported that slightly less than half of the forty neuroblastomas occurred in the children under three years of age, only five of the patients were over fifteen years old.

Ganglioneuroma may occur in either child or adult, but is more frequent in the latter. PACK and TABAH reported that ganglioneuromas may appear during early childhood, but the majority of cases do not appear until slightly later in life. Recently, SHIMADA has reported a case of retroperitoneal ganglioneuroma occurred in a 67-year-old man. There is no sex predilection for neuroblastoma or ganglioneuroma. Our case was a 22-month-old female infant.

CLINICAL FINDINGS : Neuroblastoma is rapidly growing, highly malignant tumor, and as above mentioned, it occurrs almost exclusively in infants and young

children. In this age, Wilms tumor and teratoma must be differentiated.

Ganglioneuroma is slowly growing, benign tumor. It produces symptoms, as CHRISTOPHER describes, only by pressure and pressence of a tumor.

SUMMARY

A case of ganglioneuroblastoma arising from the right lumbar sympathetic chain is presented along with a brief review of the literature pertaining to this rare tumor.

Tumor of the sympathetic nervous system is described mainly from the pathological point of view.

In closing, we wish to thank Assist. Prof. Ch. KIMURA, M. D. of our clinic and Assist. Prof. T. NISHIZUKA, M. D. of department of pathology for their valuable advice to this report. We are also indebted to Dr. R. INOUE and Dr. T. SAITO for their helpful suggestion in the preparation of this paper.

REFERENCES

1) Anderson, W. A. D.: Pathology. Mosby, 1948. 2) Boyd, W.: Text-book of pathology. Febiger, 1947. 3) Bruning, F.: Die chirurgie des Vegetativen Nervensystems. Berlin, Julius Springer, 1924. 4) Christopher, F.: A textbook of surgery. W. B. Sanders company, 1944. 5) Donhauser, J. L. et al.: Primary Retroperitoneal Tumors. Arch Surg. **71**, 234, 1955. 6) Fulton, H. et al.: Roentgen examination in retroperitoneal tumors of children. Arch. Surg. **70**, 178, 1955. 7) Fukuoka, Y.: An autopsy case of anaplastic neuroblastoma in an adult originating from the retroperitoneum. Gann no rinsyo. **2**, 70, 1956. 8) Gross, R. E.: Surgery of Infancy and Childhood, Saunders, 1941. 9) Hayashi, T.: A case of retroperitoneal ganglioneuroma. Arch, f, Jap, Chir, **23**, 114, 1954. 10) Kurata, K.: Sympathogonioma. Shikoku Acta Medica. **8**, 51, 1956. 11) Kojima, J.: Uber einen Fall von maligner Mark-geschwulst der Nebenniere. Kyoto Igaku Zassi. **23**, 200, 1926. 12) Kurobane, T.: Clinical Pathology of the Mediastinal Tumor observed from Operation Materials. Surgery, **19**, 561, 1957. 13) Lewis, D. et al.: Tumors of sympathetic nervous system. Arch. Surg. **28**, 16, 1934. 14) Mori, s.: Allgemeine Pathologie. Nihon Isyo, 1945. 15) Oda, K.: Uber einen Fall von Sympathogonioma. Okayama-Igakkai-zasshi. **43**. 1451, 1931. 16) Pack, G. et al.: Primary Retroperitoneal Tumors. Surg. Gynec. & Obst. **99**, 209, and **99**, 313, 1954. 17) Sakai, K.: Sympathicogonioma of the adrenal medulla. Arch. f. jap. Chir. **24**, 326, 1955. 18) Suzue, K.: Hypernephroma medullare. Medicine. **8**, 191, 1950. 19) Sugioka, Z,: Sympathogoniom. Arch. f. jap. Chir., **14**, 577, 1937. 20) Shimada, K.: A case of Retroperitoneal Ganglioneuroma. Surgery. **18**, 214, 1956. 21) Willis, R. A.: Pathology of Tumours. Butterworth, 1953. 22) Yamagata, Y. et al.: 4 cases of tumors of sympathetic nervous system. Gann, **46**, 225, 1955. 23) Yao, K. et al.: A case of Sympathoblastoma of an Adult woman. Gann, **46**, 231, 1955.

和文抄録

交感神経系の腫瘍
Ganglioneuroblastoma の1例

京都大学医学部外科学教室第2講座（指導：青柳安誠教授）

恒川謙吾・武田温雄・柏原貞夫

2才の女児，腹部腫脹を主訴として入院した．右側腹部に小児頭大の腫瘤を触知し，レ線的に腎外性後腹膜腫瘍である事を確めた．腫瘍の剔出手術を行い，この腫瘍は右側腰部交感神経索より発生せるものである事が判明した．

交感神経系より発生する腫瘍につき病理組織，発生頻度，部位，年令等について考察を行つた結果，我々の症例は Ganglioneuroblastoma と名附けられるものであつた．

脊髄損傷麻痺尿路炎にみた菌交代症

慶応義塾大学医学部整形外科学教室（指導：岩原寅猪教授）

武　田　　智

〔原稿受付：昭和32年10月11日〕

SPONTANEOUS OCCURENCE OF NEW BACTERIAL INFECTIONS IN THE COURSE OF TREATMENT FOR PARALYTIC CYSTITIS ON THE LESION OF SPINAL CORD

by

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Department of Orthopaedic Surgery School of Medicine, Keio University
(Director: Prof. Dr. TORAI IWAHARA)

A male patient, age of 20, was admitted in our clinic on the compressious fracture of the seventh thoracic vertebra with the lesion of spinal cord caused by falling over precipice. He was treated of cystitis due to bacterium colli that occured as complication after his hospitalization by frequent administration of mycillin, streptomycin, chloromycetin, terramycin, aureomycin and collimycin, where upon, candida was detected in his urine and he still had active cystitis.

The treatment of inflamation of the paralytic bladder due to the lesion of the spinal cord with antibiotic preparations is seemed to be no more than that of the symptome. What is important is to obtain an ideal automatism of the paralytic bladder.